# Pulmonary Heart Disease

## A Panel Discussion

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DR. ELIASER: The discussion on pulmonary heart disease will be limited to the chronic form, or what has been termed chronic cor pulmonale. For expedience we shall discuss only those conditions in which cardiac or circulatory disturbances result from diseases of the lung or the pulmonary artery or its branches.

We shall exclude acute cor pulmonale and the various forms of right ventricular failure that are frequently associated with nonpulmonary conditions such as left ventricular failure and mitral stenosis. We shall also omit those lesions characterized by left to right shunts—namely, congenital patent ductus arteriosus, interatrial and interventricular septal defects and malformations in which the pulmonary veins empty into the right atrium or superior vena cava.

The panel will be primarily concerned with those chronic states where cor pulmonale results from either one or both of the following conditions:

1. Chronic diffuse obstructive emphysema.
2. Pulmonary vascular tree lesions—intraluminal or extraluminal and in combination.

The first thing that I would like to ask the panel, which, incidentally has representatives from both Northern and Southern California, is, "Has the incidence of pulmonary heart disease increased, and, if so, why?"

### INCIDENCE

DR. PEARCE: The incidence of pulmonary heart disease is very definitely on the increase. Institutions that publish the incidence of various types of heart disease have shown a pronounced increase in this disease. Question arises as to whether this is because we now recognize it and didn't before, or is it really increasing. In my mind, there is no doubt at all that there is a decided increase in pulmonary heart disease and I suspect that as we keep people alive longer with chronic lung disease, as we do more thoracic operations, we are going to continue to see even more pulmonary heart disease.

DR. MOTLEY: That's my feeling also—that people are being kept alive longer today with better treatment both medically and surgically, and with the antibiotics life is prolonged until pulmonary heart disease may show up.

DR. ELIASER: These gentlemen, obviously, are from Southern California. Dr. Selzer, do you think it has anything to do with the air we breathe?

DR. SELZER: It may, but I would like to take the opposite standpoint in part. I think that the diagnostic facility and the accuracy with which we diagnose cor pulmonale now plays a part in the statistical increases; and my own observation has been that there may be certain overdiagnosis and we have on many occasions suspected cor pulmonale in patients with emphysema just to find out that they had the ordinary variety of coronary disease which happened to occur in patients with emphysema. It may be that the pendulum swung a little too far to the other side.

DR. MOTLEY: Could I interrupt just one second? I'd be interested to know what the feeling of the other members of the panel is about smoking.

DR. ELIASER: Do we have any volunteers?

DR. PEARCE: We're studying that aspect, but we have not been able as yet to show any quantitative difference from the standpoint of pulmonary function, either as to smoking or smog.

DR. ELIASER: We might go on to the classification of the various lung diseases that result in pulmonary heart disease. I think we agree that chronic diffuse destructive emphysema ranks No. 1 on our list. I would like to know if anyone on the panel would like to discuss the various causes of diffuse emphysema, including chronic bronchitis.

DR. PEARCE: As to the causes of diffuse obstructive emphysema: In almost all instances persons in whom the condition ultimately develops have a

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prolonged history of repeated pulmonary infections, usually with chronic bronchitis that flares up and subsides and may be associated ultimately with bronchiectasis. It is rather unusual to see a patient with late stages of a pure obstructive emphysema; there is always a degree of pulmonary fibrosis of restrictive aspect to the pulmonary disease as well as the obstructive aspect. Because of the important things we can do in prevention, it should be remembered that, almost always, obstructive emphysema is the late result of chronic pulmonary infection, with or without asthma.

DR. SELZER: I would agree with that. I think we probably should draw a very distinct dividing line between obstructive emphysema and the ordinary variety of senile emphysema (which some observers even refuse to call emphysema, using terms like senile kyphosis instead) in which the effect on the pulmonary function is relatively slight and the effect on the circulatory system is practically nil.

DR. MOTLEY: I'm one who feels that emphysema is not necessarily a part of the aging process. Our studies indicate that normally even in persons up to 70 years of age and beyond, the residual air is not above 35 per cent of total lung capacity. I know that there are some investigators who will disagree with this figure. Comroe in his book on the lung gives a figure as high as 50 per cent with no symptoms referable to the cardiopulmonary system. This is contrary to my experience. I feel the selection of cases for determining the upper limits of normal cannot be made alone from the x-ray studies or the subject's story, and that the clinical selection of what is considered normal may be misleading. If two criteria are used, (1) a normal maximal breathing capacity; and (2) a normal timed vital capacity, regardless of age, in my experience the residual air has not been found above 35 per cent of total lung capacity, so I don't feel that emphysema is part and parcel of the aging process normally. There are many persons with a residual air above 35 per cent of total lung capacity who are capable of doing their jobs and have no complaints. The reason for this is the big reserve in the lung normally, so that by the time subjective complaints of dyspnea are present the lung changes represent an advanced stage. I feel that residual air above 35 per cent of total lung capacity is abnormal, just the same as systolic blood pressure of 200 mm. of mercury is abnormal even though the individual may have no subjective complaints. Infection and bronchospasm are two very important etiologic factors in pulmonary emphysema. Also, there appears to be a hereditary factor—a difference in protoplasm, making some persons more prone to the stresses and strains that cause emphysema to develop. Smoking also appears to be an important factor in emphysema in some cases, although difficult to prove, for many heavy smokers never develop emphysema. It is of interest that practically all the patients I have observed with severe emphysema who have never been exposed to any kind of dust inhalation such as might produce emphysema (as silicate, diatomaceous earth or asbestos) do have a history of prolonged heavy cigarette smoking—two or three packs a day.

DR. ELIASER: Regarding the second major classification of pulmonary diseases, I should like to ask the panel what are the more common varieties of pulmonary vascular diseases that produce this condition, and what do we mean by alveolar capillary block?

### ALVEOLAR CAPILLARY BLOCK

DR. SELZER: I think that alveolar capillary block simply means that there is difficulty in diffusion between the capillaries and the alveoli. I don't know whether we can consider this actually a circulatory disease. I think it is a disturbance in the gas exchange which does not necessarily lead to the condition that we are interested in here. I think perhaps one ought to state that the diagnosis of cor pulmonale or pulmonary heart disease is one of increased pressure in the pulmonary artery; in other words, what we are looking for is the effect of pulmonary hypertension on the heart, leading to its failure. We don't know what the relationship is between alveolar capillary block and pulmonary hypertension. I think perhaps we ought to separate the pulmonary function aspect of these diseases from the circulatory effect. They very often occur together but they also may be separate.

DR. MOTLEY: We have been interested in trying to get more precise information as to why the oxygen saturation of arterial blood is reduced in emphysema. When special tests were used, there appeared to be little evidence of a true diffusion difficulty at the alveolar-capillary membrane, such as typically occurs in beryllosis—which is a classic example of diffusion difficulty. The common finding in emphysema demonstrated every day is the presence of poorly ventilated alveoli, and with exercise most often a drop in saturation occurs, due principally to the shunting of blood through nonventilated areas or very poorly ventilated areas.

Two very simple tests can be used to demonstrate these findings. In the past few years a great deal has been written about the diffusion capacity of the lung. Reduced diffusion capacity of the lung is a general term and the condition may be due to several things and does not necessarily involve a true diffusion difficulty at the alveolar-capillary membrane with an increased arteriovenous gradient for oxygen

—for example, a reduced blood flow in pulmonary stenosis or in pulmonary hypertension with septal defect. The two simple tests entail high oxygen breathing and pressure breathing on air. In one the inspired oxygen tension is increased on a 32 per cent oxygen breathing mixture enough to overcome a diffusion difficulty, if that is the primary difficulty on air breathing. The decrease in saturation with moderate exercise, which occurs most often in emphysema, is not overcome—that is, saturation does not return to normal—even on the high oxygen mixture (32 per cent oxygen), thus demonstrating the absence of diffusion difficulty. The other test is the use of intermittent positive pressure breathing with compressed air only—no bronchodilators. In most cases of emphysema a significant rise in the saturation, sometimes up to normal, results from the pressure breathing on compressed air only; and the only way to explain this finding is on a basis of the presence of poorly ventilated alveoli, with the pressure breathing providing improved aeration.

Persons with emphysema have impaired movement of the diaphragm, loss in lung elasticity and increased breathing resistance, so that alveolar aeration is impaired and constitutes an important aspect in the blood gas exchange. If the saturation drops with exercise in a patient with emphysema, the low oxygen saturation represents a period of acute hypoxia. Acute hypoxia elevates pulmonary artery pressure and increases resistance. If the saturation drops 5 per cent or 10 per cent, the patient is advised not to walk up steps or uphill any more than he has to—or, if he lives on a second floor apartment, to get a first floor apartment. Every little acute episode represents an insult with a little increase in work load on the right side of the heart, and hastens the day right heart failure develops.

DR. PEARCE: My impressions are pretty much those which have been mentioned here—that is, that the primary difficulty leading to pulmonary hypertension in patients with cor pulmonale is basically one of distribution of gas to the right place and that alveolar capillary block is a relatively rare thing.

#### POLYCYTHEMIA

DR. ELIASER: The next thing we should discuss is the occurrence of polycythemia in pulmonary heart disease. Is it of any prognostic importance and does it help in differentiating between patients with considerable emphysema and those with little?

DR. PEARCE: This is an area in which I think you get many different impressions. Mine is that patients who get polycythemia fairly early in the course of pulmonary disease are the patients who tend to get cor pulmonale. Of course, by and large, they are the people with the most severe anoxia in the obstructive group, also the patients who have the most severe $CO_2$ retention.

DR. MOTLEY: My co-workers and I have been unable to make any correlation whatsoever between the hemoglobin values and the severity of pulmonary emphysema as measured by function measurements. If the hemoglobin is elevated we suspect a cardiac factor in emphysema. The oxygen capacity method, which I think is one of the more accurate measurements, is used to determine hemoglobin, and all of our experience has been at sea level pressure. There appear to be some differences between sea level and 5,000 feet altitudes like Denver and Salt Lake City, according to Dr. Hecht. Although there is no correlation between the severity of the emphysema and the hemoglobin level, if the patient starts going into the slightest degree of right heart failure, the hemoglobin level goes up. Hence, increased hemoglobin indicates the presence of some element of right heart failure.

DR. SELZER: It has been my impression that the relation between pulmonary heart disease, specifically pulmonary hypertension, and polycythemia is rather vague. There are enough cases of severe pulmonary hypertension other than emphysema in which there is no polycythemia; especially, many of the patients with so-called primary pulmonary hypertensions never get polycythemia and anoxemia. On the other hand, in dealing with emphysema, hypoxia is one of the factors in the production of cor pulmonale and is also a stimulus for the formation of excess hemoglobin, so that the relationship works in one direction but not in the other.

#### RESPIRATORY ACIDOSIS

DR. ELIASER: I should now like to bring up the rather interesting problem of respiratory acidosis. Is it of any significance in the differential diagnosis between the various forms of pulmonary disease, what are the conditions that produce it, and are there any factors that tend to accelerate its onset?

DR. PEARCE: Respiratory acidosis is the thing we all fear particularly in these cases, because it is at this point, when respiratory acidosis becomes severe, that the end is usually in sight unless the cause is something that is quickly correctable, such as a pulmonary infection. Respiratory acidosis is seen most commonly in the obstructive diseases. In those in which there is primarily an abnormality in the distribution of gases, anoxia occurs long before carbon dioxide retention does, because $CO_2$ as a gas is about 20 times more diffusable than oxygen.

DR. MOTLEY: Respiratory acidosis develops when alveolar ventilation is inadequate to blow off the carbon dioxide. The body is giving off almost as much

$CO_2$ all the time as oxygen is taken in, and, as Dr. Pearce has mentioned, the $CO_2$ in solution is about 25 to 30 times as diffusable as oxygen across the pulmonary membranes. Hence, for all practical purposes the $CO_2$ in the arterial blood is the same as in the alveoli—that is, the mean $pCO_2$* pressure. The determination of the arterial $pCO_2$ provides a measure of the adequacy of alveolar ventilation. If the arterial $CO_2$ content or $pCO_2$ is elevated, then alveolar ventilation is inadequate. However, one cannot determine from the $CO_2$ level whether the patient is in acidosis or not at the time. Many patients with severe emphysema always have high $CO_2$ levels. The $pCO_2$ may be 60 mm. of mercury or more (normal 40 mm. at sea level), yet the arterial blood pH be 7.45. In other words, they are compensated. The emphysema has developed over a period of years and they are compensated to the increased level. Trouble develops when such a person gets an infection with bronchospasm and plugging with mucus and secretions. The alveolar ventilation is reduced still more in the presence of little or no reserve and immediately the $CO_2$ goes up still more and the pH goes down below 7.38—that is, the arterial blood pH, which is the pH we must measure to have a guide of what is going on in the lung (venous blood is unsatisfactory).

The patient in acidosis requires prompt treatment to increase alveolar ventilation. The arterial blood pH is a real and indispensable guide in determining whether or not acidosis is present in these people with severe degree of pulmonary insufficiency. In my experience the calculated $pCO_2$ values or the calculated pH values in chronic pulmonary insufficiency of severe degree are not accurate even when one knows the arterial $CO_2$ content; and the $CO_2$ combining power may be completely misleading.

#### CARDIAC OUTPUT

DR. ELIASER: Now I have a more difficult question. Why is it that some persons with pulmonary heart disease have ostensibly high cardiac output and some have low output?

DR. SELZER: We have been interested in this subject and have studied the cardiac output in a series of between 20 and 30 patients with cor pulmonale of various degrees of severity. In our experience the high output is very rare. We have had, I think, only one or two patients in whom the cardiac output was definitely abnormally high. There are some in whom perhaps in relation to their symptoms, the output appears to be higher than average but is still within the normal zone. I think most of these people sooner or later as they develop more circulatory disturbance go into low output failure—that has been our experience.

DR. MOTLEY: I would agree with that with regard to the resting values, and certainly with exercise they are unable to increase the cardiac output normally corresponding to the exercise given. Some patients almost seem to have a fixed level to which the output can be increased, only a few liters above the resting level, regardless of how much exercise is given. With the more severe exercise on a treadmill or bicycle, they'll just fatigue quicker. One simple test which reflects the ability to increase the pulmonary blood flow is the exercise oxygen uptake. If the exercise oxygen uptake does not increase to a degree corresponding to the degree of exercise given, and if the minute ventilation is within the normal range, this finding indicates that the pulmonary blood flow is reduced, and indirectly that pulmonary vascular resistance is increased.

DR. PEARCE: We have been looking for patients with high output failure, so-called, due to pulmonary disease, and we certainly have been able to find very few. We've done dilution output studies on a large number of patients with chronic lung disease looking particularly for this group. I think the point Dr. Motley makes is a very good one—that even when you do find patients with moderately elevated output (and we've found them even as high as 10 liters per minute) with exercise they are not able to raise their output at all.

#### CLINICAL ASPECTS

DR. ELIASER: We might now comment on some of the clinical aspects of this condition and inquire whether there are any bedside clues to the presence of pulmonary heart disease in either the patients' symptoms or in the detection of any specific physical signs.

DR. PEARCE: Well, as far as determining cor pulmonale in a sense of hypertrophied right ventricle in itself is concerned, I think this is difficult to do by clinical signs alone. Certainly one can be suspicious about it when one finds the pulmonary disease which one can commonly detect by physical signs, particularly obstruction and restriction of ventilation. I think that as far as the physical signs of right hypertrophy itself go, looking for the accentuation of the second pulmonic sound, as has been mentioned earlier here today, is not particularly helpful. I have found that the appearance of a diastolic gallop along the sternal border has been a more useful sign than the accentuation of the second pulmonic sound which, in patients with barrel chests particularly, is something you may never hear at all, even with an extraordinarily high pulmonary artery pressure.

*Abbreviation for partial carbon dioxide.

DR. SELZER: I would agree with Dr. Pearce's statement. I think if the chest is not too barrel-shaped, we can also pay some attention to the right ventricular type of pulsation, which is quite helpful in diagnosing right ventricular hypertrophy.

DR. PEARCE: Sometimes when you can't detect this pulsation along the left sternal border because of a large chest, you can sometimes feel it by hooking your fingers under the xyphoid process and then feel a very active right ventricle.

DR. MOTLEY: I think I might add that one should be on the lookout for pulmonary heart disease in a patient where the breath sounds are distant. The evaluation with the stethoscope and the physical examination of the chest is better correlated with the degree of emphysema by pulmonary function measurements than by x-ray films. The use of the stethoscope is definitely indicated in chronic pulmonary disease, although some have advocated throwing it away. So often the patient comes in with right heart failure, and the cardiologist treats the right heart failure, but sometimes overlooks the underlying pulmonary disease.

#### SPECIALIZED DIAGNOSTIC PROCEDURES

DR. ELIASER: We can now move on to some of the more specialized diagnostic procedures; for example, what are the indications for pulmonary function studies and which ones are more frequently used?

DR. MOTLEY: I think any case that presents the clinician with a pulmonary problem in which there is possibility that function may be impaired, demands a little further investigation. Spirograms offer a rather simple test that can be done very quickly. The information obtained consists of total vital capacity, timed vital capacity, and the maximal breathing capacity, with a further determination of the maximal breathing capacity after administration of a bronchodilating drug (a test for bronchospasm). Also, a record of the shape of the spirogram tracing is very desirable, because in emphysema there is a very characteristic pattern with prolongation of exhalations (in some instances up to 12 to 15 and 20 seconds or more). One can often get some very useful information by asking the patient to take a deep breath and then to blow out as rapidly as possible. If the blow out is prolonged for 10 to 15 seconds, the extra time is very readily apparent; some patients even wheeze. Total vital capacity should never be used alone as a single measurement, as it may be misleading in some instances. The spirogram measurements may be indeterminate in some cases, or if the readings are either normal or extremely abnormal, that may be all that one needs in a particular case. If the spirogram measurements are moderately reduced, then we need to measure residual air to evaluate the severity of the emphysema. Residual air measurement is the only way that we have for accurately assessing quantitatively the degree of pulmonary emphysema.

Other tests, such as the use of a nitrogen meter with a single deep breath of oxygen or measurement of the nitrogen washout by individual breaths, require a complicated electrical set-up. The residual air is a simple measurement, but it is a laboratory and not an office procedure.

To complete the study, the blood exchange must be evaluated. Even though the patient may not have cyanosis, he may still have unsaturation and one must compare rest and exercise. Hemodynamically, rest and exercise represent two different situations in a given patient. The resting saturation may be almost normal, but exercise will bring out gross abnormalities. Arterial blood studies may be all that are needed in some cases. Exercise oxygen uptake has already been referred to as a very useful test and well correlated with complete function measurements where the pulmonary blood flow is reduced regardless of the cause, whether it be pulmonary emphysema or pulmonary hypertension with an atrial septal defect or with pulmonary stenosis. Anything that reduces the pulmonary blood flow will reduce oxygen uptake. Every case presents a specific problem, and in some just a few tests such as are provided by spirograms are in themselves adequate, or they may point the need for more complete study. I know of some physicians who have the Collins 13.5 liter respirometer in their office (probably the best apparatus for obtaining this information). I know of one chest physician who takes a spirogram on practically every patient who is getting an x-ray film of the chest.

DR. SELZER: I have nothing to add to the pulmonary function studies. I think, however, that it should be emphasized that in cor pulmonale there is no other way of actually knowing what one is dealing with than by cardiac catheterization. Pulmonary artery pressure at rest and exercise cannot be judged by any other indirect means.

DR. PEARCE: The only thing I have to add to what has been said is that in the very ill patient where the tests of respiratory function are very difficult to do, when we have acute situations in the hospital we find that frequent determination of the pH and the $CO_2$ content of the arterial blood gives the most help in determining which way they are going. I might say here parenthetically to beware of the report of $CO_2$ combining power on venous blood which has been suggested by some observers to be a good measure of $CO_2$ retention. This is a real booby trap, I think.

## TREATMENT

DR. ELIASER: Now, as to treatment. I should like to bring up one question. There was a concept several years ago that digitalis may be bad for patients with pulmonary heart disease or that it may not do very much good. I'd like to get an expression of opinion from the panel: Should patients with pulmonary heart disease who are in congestive failure receive digitalis and, if so, which one of the glycosides?

DR. PEARCE: I think they should by all means receive the glycosides and I'm not particularly partial as to which they receive. Unless I'm in a hurry, I generally use digitalis leaf. Some of the bad results in the use of glycosides in these patients have been due to treating only the myocardium and not treating the pulmonary infection and obstruction in the patient with severe emphysema who is in failure.

DR. SELZER: Dr. Eliaser, I think that the concept that you mentioned originated in England; I think they may have changed their views since, and they all agree that digitalis is not contraindicated. However, I think it is very strongly disappointing in cases of cor pulmonale. When we use it, and I think we should, we very seldom see any spectacular effect.

DR. ELIASER: Another controversial therapeutic measure is phlebotomy. When should that procedure be used, and what should be used as a criterion for discontinuing it?

DR. SELZER: This is a subject of considerable controversy, for polycythemia increases the oxygen-carrying capacity and in a way is considered a useful compensatory measure. Most physicians feel, however, that it overshoots usefulness and may become harmful. One should try not to expect too much of phlebotomy in moderate polycythemia, but when the packed cell volume is 60 per cent or more, bleeding should be done to bring it down to about 55 per cent.

DR. ELIASER: I'm about to carry coals to Newcastle. Dr. Motley, when is intermittent positive pressure breathing indicated and what are its contraindications, if any?

DR. MOTLEY: I feel that intermittent positive pressure breathing (IPPB) has a very definite place in the treatment of chronic pulmonary disease. In severe emphysema IPPB is the most efficient method for the administration of bronchodilators and antibiotic, if the patient has an infection in which an antibiotic is indicated. The main function of the lung is to get oxygen into the blood and get rid of $CO_2$ adequately, and physiological studies indicate that IPPB improves this function in emphysema. Secondly, intermittent positive pressure breathing promotes bronchial drainage and helps raise secretions. This has been observed especially in coal miners as a group. Coal miners who have been away from the mines for as long as two years have started spitting black soon after IPPB was started. The third aspect with IPPB is breathing exercises. A lot has been written on breathing exercise, but in my experience it is hard to get patients to systematically take breathing exercises that are effective, and often in severe emphysema they cannot take effective breathing exercises. A patient with emphysema may be fluoroscopically observed to have very little movement of the diaphragm and a shallow type of breathing, but when fluoroscopy is repeated with IPPB increased movement of the diaphragm and deeper ventilation may be seen; and this is reflected in the arterial blood studies, for the oxygen tension goes up and the $CO_2$ goes down with administration of compressed air only (no oxygen with bronchodilators). The net result of IPPB is to keep the airways open by more effective treatment of bronchospasm and promoting bronchial drainage.

Follow-up studies, after three years, of patients with severe emphysema who have their own IPPB units for use at home and who have taken the treatments regularly and properly have shown that the residual air was not increased. I think that the only way patients with severe emphysema can hope to obtain the maximum benefits possible from the use of IPPB is to have a unit for use at home where, to start with, they may take three or four treatments a day of 15 to 20 minutes each. Eventually the number of treatments may be cut down to one or two a day in the less severe cases. However, the patients are going to require treatments for emphysema for the rest of their lives, as this condition tends to be slowly progressive; and unless one can prevent progression of the increased residual air (the best method to evaluate changes over long periods) the course is progressively downhill. Patients with emphysema often date the beginning of illness from an infection which calls it to their attention, although the emphysema has been present for many years, slowly progressive. Intermittent positive pressure breathing should be used in conjunction with all the other treatments indicated, such as use of diuretics, digitalis, low sodium diet, antibiotics and wetting agents. IPPB can be used in conjunction with tracheotomy, and in a few cases this has been life saving. IPPB can be used to supplement the Drinker respirator, especially with tracheotomy. The emphysema patient gets an infection, bronchospasm and the smaller bronchioles become plugged, alveolar ventilation is decreased, $CO_2$ increases, and respiratory acidosis develops; death then follows unless something is done about improving alveolar ventilation, and supplementary pressure breathing may be required continuously for several days.

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